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Original Research Paper

General Surgery

RETROPERITONEAL NON FUNCTIONING PARAGANGLIOMA- A CASE REPORT

Dr Mahesh K Professor, Department of General Surgery, JJMMC, Davangere Karnataka

Dr Jagadeesha BVC* Professor, Department of General Surgery, JJMMC, Davangere Karnataka
*Corresponding Author

Dr Mahesh Polamreddy Post Graduate, Department of General Surgery, JJMMC, Davangere Karnataka

ABSTRACT Paragangliomas are extra adrenal neoplasm arising from sympathetic or Parasympathetic ganglia and are derived from chromaffin cells. These are rare asymptomatic and pre operative diagnosis is seldom common. We report a case of such retro peritoneal paraganglioma in a 35 year old presented with vague pain abdomen and normal serum catecholamines. Complete resection of tumour done and HPE confirmed paraganglioma.

KEYWORDS : paraganglioma NET,organ of zuckerkandl

INTRODUCTION:

Paraganglioma are one of the rare neoplasms arising form neural crest cells similar to neuroblastoma, ganglioneuroma,Crotid body tumour. Extra adrenal phechromacytomas are considered as paraganglioma usually arise from organ of zuckercandl and can also be seen in mediastinum,skull base and neck, Functional tumour secrete excess catecholamines and hence they are symptomatic. Symptoms include headache, sweating and palpitations with episodic or persistent hypertension as an important sign.10-15 % of the are non functional and pose significant diagnostic challenge of this reports. We describe a new case of non functional retroperitoneal paraganglioma.

Case presentation:

A 35 year old male patient presented with vague abdominal pain of 1 month duration with similar multiple episodes in past 6 months with no h/o headache,sweating and palpitations. He is not a known case of hypertension and was not on any treatment. No family history of similar illness noted. Physical examination reveals a retroperitoneal mass of size 7x8 cm in the umbilical, left iliac fossa, hypogastrium and left lumbar region. Mass was immobile and was not prominent in knee elbow position. USG shows large retroperitoneal mass para aortic plane with indentation in IVC MDCT Showed large lobulated heterogenous enhancing soft tissue mass with central necrotic focus in the mid abdomen abutting the jejuna loop and infra renal aorta on the right side noticed. IVC is partially compressed. Perilesional fat planes are fairly preserved. Lesion measures about 6.7 x 7.5 x 6 cms with perilesional small lymph nodes.

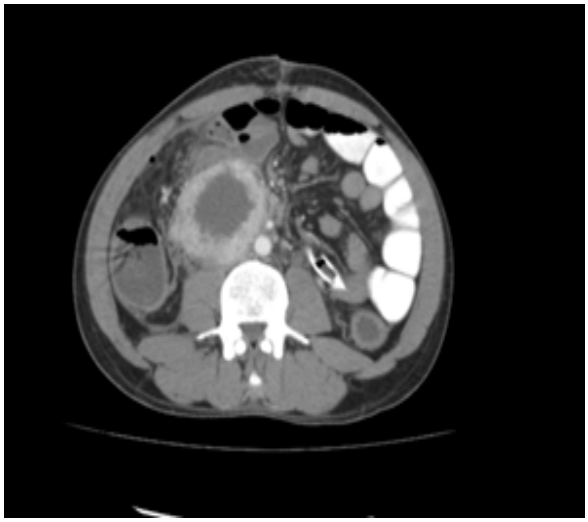

FIG 1:MDCT cross sectional image

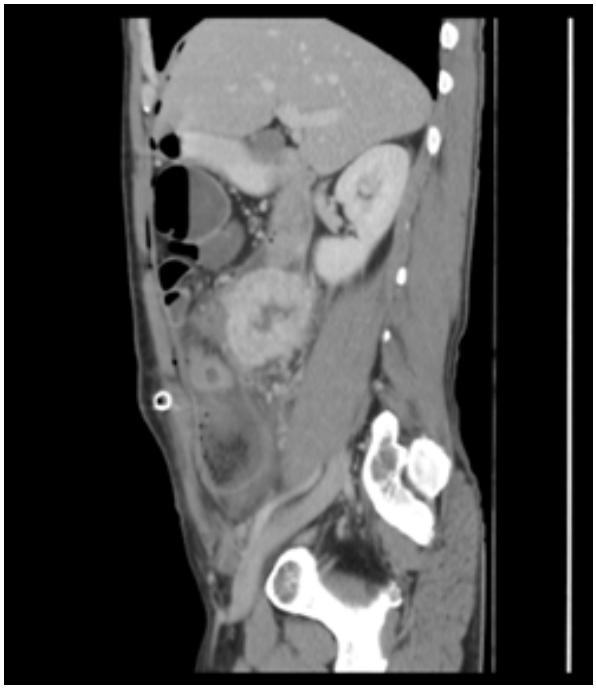

FIG 2: MDCT sagittal section image

The lesion was completely resected with midline laparotomy.on macroscopic examination tumour of size 6x8.5x5 cm as noted with grayish pink on the cut surface and rich vascularity. Are of haemorragic and necrosis were observed.

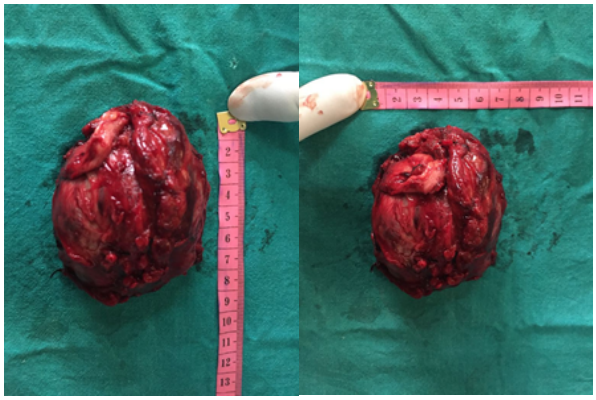

FIG 3: measurements of specimen

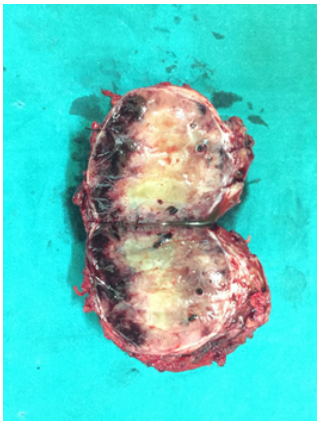

FIG 4: cut section of specimen

Histopathological examination identified circumscribed lesion with fibrous capsule predominantly arranged in nestswith fibrovascular septae forming zellballen pattern. Tumour cells are predominantly monotonous with few gaint atypical cells having moderate granular eosinophilic amphophillic cytoplasm with areas of heamorrhage. All these features suggestive of benign paraganglioma

DISCUSSION:

Pheochromocytoma and extra-adrenal paraganglioma are tumours derived from catecholamine-producing chromaffin cells of neural crest origin with incidence of 3-8 per million per year. 10-15% of them are asymptomatic and pose diagnostic challenge. 0.05% of all autopsies have these tumors[1]. Traditionally termed '10% tumour'(10% bilateral, extra-adrenal,familial or malignant), this description has now been challenged by recent advances in genetics and diagnosis[2]. Majority of paragangliomas occur in abdomen in organ of zuckerkanndl. In contrast to sympathetic paragangliomas those occur in head and neck tend to be arising from parasympathetic ganglia which rarely produce catecholaminnes. One third of these are associated with syndromes like MEN syndrome, von Hippel-Lindau syndrome and neurofibromatosis type-1. Mutations occur in SDH gene encoding subunits of succinate hydrogenase enzyme responsible of 'paraganglioma pheochromocytoma syndrome'[3].

The classical triad of presentation in headache, palpitations and sweating associated with episodic hypertension. Other symptoms vague abdominal pain, nausea, tiredness, loss of weight, anxiety. Postural changes, exercise and anxiety may provoke symptoms. Patient can be normal in between the episodes and key to diagnosis is stay alert to the possibility of the disease. Plasma free or urinary fractionated metanephrines are now established as the best diagnostic test for pheochromocytoma[4-6]. Their levels correlate with the tumor mass. In case of classical symptoms MIBG scan is investigation of choice. To plan for surgery MRI of the the site of interest has better value than CT scan[7]. PET scan has a role metastatic disease. Metastatic disease is rare and associated with younger patients with syndromes and large tumors (>6cms).

Laparoscopic adrenalectomy is the procedure of choice for resection of tumor. Open approach is preferable for larger tumors (>6cms)[8]. Before planning for surgery effective control of hypertension using phenoxybenzamine(started 20mg BD with 10mg increments till postural hypotension is reached) is a must to avoid catastrophic blood pressure changes intra operatively.

REFERENCES

1. Eisenhofer G, Bornstein SR, Brouwers FM, et al. Malignant pheochromocytoma: current status and initiatives for future progress. Endocr Relat Cancer 2004; 11(3):423–36.
2. Elder EE, Elder G, Larsson C. Pheochromocytoma and functional paraganglioma syndrome: no longer the 10% tumor. J Surg Oncol 2005;89(3): 193–201.
3. Gimenez-Roqueplo AP, Lehnert H, Mannelli M, et al. Phaeochromocytoma, new genes and screening strategies. Clin Endocrinol (Oxf) 2006;65(6):699–705.
4. Grossman A, Pacak K, Sawka A, et al. Biochemical diagnosis and localization of pheochromocytoma: can we reach a consensus? Ann N Y Acad Sci 2006;1073:332–47.
5. Lenders JW, Pacak K, Walther MM, et al. Biochemical diagnosis of pheochromocytoma: which test is best? JAMA 2002;287(11):1427–34.
6. Pacak K, Eisenhofer G, Ahlman H, et al. Pheochromocytoma: recommendations for clinical practice from the First International Symposium, October 2005. Nat Clin Pract Endocrinol Metab 2007;3(2):92–102.
7. Ilias I, Pacak K. Current approaches and recommended algorithm for the diagnostic localization of pheochromocytoma. J Clin Endocrinol Metab 2004;89(2):479–91.
8. Shen WT, Sturgeon C, Clark OH, et al. Should pheochromocytoma size influence surgical approach? A comparison of 90 malignant and 60 benign pheochromocytomas. Surgery 2004;136(6): 1129–37.